Complete congenital heart block
Report of 35 cases

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SUMMARY Congenital complete atrioventricular block is uncommon, and the outlook is usually regarded as favourable. Thirty-five patients with congenital heart block are presented. There was no obvious sex difference and their ages ranged from 12 days to 85 years, though most were under the age of 20 years when first seen. Accompanying heart disease was noted in six, but presentation with symptoms in early infancy was a more serious risk factor in our experience. Permanent pacing was required in 21, three of whom were neonates. One of the 14 unpaced patients died unexpectedly at the age of 5 years. Long term supervision is necessary, as most will require permanent pacing before their 50th birthday.

Acquired third degree atrioventricular heart block in adults is relatively common, being attributed most frequently to degeneration of the fibres in the conducting system.[1] Congenital complete heart block, by contrast, is rare. It was first described by Morquio,[2] who reported a family in which several sibs had pronounced bradycardia and syncopal attacks, with death in early childhood. We encountered 35 patients with congenital heart block, and the purpose of our paper is to try and assess the incidence of the condition in our region, the possible prognosis, and from the study to define the indications for establishing permanent pacing.

Patients and methods

Over a period of 18 years (1962 to the end of 1980) we encountered in our region in south-west Scotland a total of 35 patients with congenital complete atrioventricular block. All fulfilled the criteria defined by Yater[3]: a slow heart known to have been present from birth or from a very early age; the absence of a history of diphtheria or other myocarditis which might cause heart block; no evidence of ischaemic heart disease or cardiomyopathy; no previous cardiac surgery.

Fifteen of the patients were male and 20 female. The age at presentation is tabulated (Table 1). The youngest patient was 12 days old and the oldest was a man of 85 years known to have had a slow heart rate since early childhood and who was subsequently discharged from military service in the first world war on this account. He remained symptom free until his mid-eighties when he developed syncopal attacks and was referred for permanent pacing.

Of the four who presented in early infancy, three had symptoms and required early, permanent pacing, while the fourth who had no symptoms was discovered to have bradycardia on routine medical examination. The remaining patients were referred in later childhood or early adult life because of the finding of a slow heart rate, with or without symptoms. Long term supervision was considered mandatory in all. Twenty-one patients required the insertion of a permanent pacemaker and they are considered separately from, and compared with, the 14 who were not paced.

ACCOMPANYING CARDIAC DEFECTS
Six of our patients (17%) had accompanying congenital cardiac defects. These comprised three with ventricular septal defect, two with persistent ductus arteriosus, and one with coarctation of the aorta with

Table 1 Age in years at presentation of 35 patients with congenital heart block

0–11/12	1–4	5–9	10–19	20–29	>30
4	2	8	13	5	3

persistent ductus arteriosus. In the remaining 29 congenital heart block was the sole anomaly.

Results

PERMANENT PACING

Permanent pacemakers were inserted into 21 patients because of symptoms comprising either syncopal attacks or signs of early cardiac failure. Only one had an accompanying cardiac defect, an infant of 3 weeks old in cardiac failure with coarctation of aorta. This was repaired surgically and a pacemaker implanted as an emergency procedure. The age at which pacing was established is set out in Table 2.

The three youngest patients were aged 12, 21, and 22 days. Emergency pacing was required because of frequent syncopal attacks in two and of advanced cardiac failure in one. At thoracotomy the pacing leads were anchored to the external surface of the heart and attached to a generator embedded either in the costophrenic sulcus or subcutaneously in the anterior abdominal wall. A small mercury chloride Medtronic pacemaker with a rate set at 120 a minute was used and replaced at two years and again at five years by a larger lithium generator.

There were no deaths or complications in the pacemaker group. All have been followed up and have remained well one to 12 years since the start of pacing.

PATIENTS NOT PACED

Fourteen patients did not undergo permanent pacing. One of these, a 5 year old boy, died quite suddenly and unexpectedly at home after a short illness accompanied by diarrhoea and vomiting. He had been free from symptoms and had no obvious accompanying cardiac defect. The electrocardiogram had shown complete heart block, with a resting heart rate of 47 and a normal QRS and QT interval. No obvious cause of death was found at necropsy.

Only one patient developed symptoms, a girl who at the age of 18 years experienced several mild syncopal attacks. In view of the unreliability of pacemakers at the time (1963), treatment with sustained release isoprenaline was started as an alternative. The attacks became less troublesome and eventually disappeared. Subsequently she had three uneventful pregnancies, and now at the age of 36 years is well with only mild dyspnoea. The resting heart remained unaltered at 44 a minute.

Table 2 *Age in years at pacemaker implantation in 21 patients with congenital heart block*

0–11/12	1–4	5–9	10–19	20–29	30–49	>50
3	0	0	5	2	9	2

Table 3 *Present age in years of 13 unpaced patients with congenital heart block*

0–11/12	1–4	5–9	10–19	20–29	30–39
1	1	2	3	5	1

The remaining 12 in this group were referred in late infancy, later childhood, or adult life because a slow heart rate had been found on routine examination. All remained well. The current age of the surviving non-paced patients is as shown (Table 3).

ANALYSIS OF ELECTROCARDIOGRAM AND HEART RATE

The electrocardiograms of all the patients were closely studied to find out whether there were any differences between those who required pacing and those who did not. The heart rate varied from 38 to 60, but in no patient did the QRS complex exceed 0·12 second. The QT interval measured 0·48 in one patient only, an infant of 21 days who required emergency pacing. In the remaining 34 the QT interval was less than 0·43 second.

ATTEMPT AT ASSESSING PROGNOSIS IN CONGENITAL HEART BLOCK

Emergency pacing was necessary in three out of four who presented in early infancy (Tables 1 and 2). The remaining 31 were first seen after the age of 1 year and none required a pacemaker until the second decade of life. Of these 31, 18 eventually required pacemakers, most of them between 10 and 49 years. Only two patients survived beyond their 50th birthday without the need for permanent pacing.

Discussion

In the absence of any reliable figures, it is difficult to define precisely the incidence of congenital complete heart block in the community. It is relatively uncommon compared with the incidence of acquired heart block in adults. Keith and his associates at Toronto Children's Hospital[4] found 90 cases of congenital heart block out of 15 104 patients with congenital heart disease between 1950 and 1973. In an international study from 44 paediatric centres in Europe, the United States, Canada, and Mexico[5] a total of 599 infants and children with congenital heart block was collected. It was estimated from this study that the incidence lay between one in 15 000 and one in 20 000 live births. The annual birth rate for south-west Scotland (population approximately three million) for the last few years has been 40 000 which would produce two cases of congenital heart block each year. In 18 years (the period under review in our study) the figure would be 36, which corresponds very closely with the

total number we encountered.

Several histological studies have been reported in congenital heart block in an attempt to localise the site of block and its cause.[6-8] In a further study on post-mortem specimens the presence of aberrant fibrous tissue either at the level of the atrioventricular node or the atrioventricular bundle was demonstrated histologically.[9] The conclusion was that this tissue interrupted normal conduction pathways, producing discontinuity between the atria and atrioventricular node or between the node and ventricular conducting tissues. His bundle studies may help to localise the site of the block.[10 11] In congenital heart block with no accompanying cardiac lesion this is situated most frequently proximal to the bundle of His.[12] We did not carry out any His bundle recordings in any of our patients.

Recently several observers reported an association between congenital heart block in newborn infants with maternal connective tissue disease, particularly systemic lupus erythematosus.[13-16] We were unable to trace any such relation in any of our patients on clinical grounds, though laboratory evidence was not available in all of them.

The prognosis in congenital heart disease particularly when there is no accompanying heart defect appears to be favourable according to reports.[4 5] It can be compatible with normal growth and development,[17] and some patients have survived to their 40's or even 60's without pacing.[18 19] In rare isolated cases, return to sinus rhythm has occurred spontaneously,[20 21] though this is more likely in heart block complicating surgical repair of congenital cardiac defects.[22-24] Successful pregnancy in patients with congenital heart block, who have not been paced, has been reported.[25] One of our unpaced patients had three uneventful pregnancies despite previous syncopal attacks.

Such a favourable prognosis is not uniform throughout childhood and early adult life in all cases of congenital heart block. Fatal Adams-Stokes attacks occasionally occur in children,[26] and one of our patients died unexpectedly at the age of 5 years, presumably from this cause. Certainly prophylactic pacing in childhood is unnecessary in most, particularly as morbidity associated with pacemakers is more likely to be the result of repeated electrode trauma and displacement of pacing leads caused by growth.[27]

In attempting to assess prognosis and the need for pacing, Moltham and his colleagues[26] suggested that a wide QRS with or without left bundle-branch block was ominous, while a QT interval exceeding 0·43 was considered a serious risk factor predicting the early onset of symptoms.[28] We did not find any significant abnormalities on the electrocardiogram in our patients, apart from a prolonged QT in one. What may be more helpful in defining which patients may require early pacing before the development of symptoms is the application of controlled exercise testing. Taylor and Godfrey[29] studied four patients with congenital heart block on a bicycle ergometer. Two were able to achieve normal maximum work even though they did so without reaching a satisfactorily increased cardiac output and with a very low oxygen content of venous blood. Winkler and his colleagues from Boston[30] subjected 25 children with complete heart block to maximum exercise testing on the treadmill using the Bruce protocol and compared the results with those of 50 normal children and young adults used as controls. Seventeen (68%) out of the 25 with heart block had significant and frequent ventricular ectopics on the electrocardiogram during exercise compared with only one (2%) out of the 50 controls. This finding they concluded could be a bad prognostic factor for possible sudden death. We did not subject any of our patients to exercise testing, but this appears to be a promising single and safe means of helping to assess the need for pacing in asymptomatic patients with congenital heart block. This is nowadays much more relevant, with the striking developments and improvements in reliability of pacemakers over the last few years.

Patients with congenital heart block presenting with symptoms in early infancy require emergency pacing. Accompanying congenital heart disease constitutes an additional risk factor. This was present in 30% of the patients in the international series reported by Michaëlsson and Engle[5] and undoubtedly affected the outcome adversely. It was found in only six (17%) of our 35 patients. Most, however, do not require pacing in childhood, but an increasing number do so from the second decade onward. Life-long supervision is imperative, as most will require a permanent pacemaker before the age of 50 years.

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